CF - Cystic Fibrosis

CF-AP ANATOMY AND PHYSIOLOGY

OUTCOME: The patient/family will understand anatomy/physiology as it relates to cystic fibrosis.

STANDARDS:

1. Discuss anatomy/physiology as it relates to cystic fibrosis and that it often is a multisystem disease.
2. Discuss changes to anatomy/physiology as a result of cystic fibrosis.
3. Discuss the impact of these changes on the patient's health or well-being.

CF-C COMPLICATIONS

OUTCOME: The patient/family will understand common and important complications of cystic fibrosis.

STANDARDS:

1. Discuss pulmonary complications of cystic fibrosis as appropriate.
2. Discuss that cystic fibrosis may affect any part of the respiratory mucosa.
3. Discuss that exocrine pancreatic failure may cause fat malabsorption and lead to growth delay or failure.
4. Discuss that endocrine pancreatic failure may lead to glucose intolerance or insufficient insulin secretion.
5. Discuss that cirrhosis may result from severe forms of cystic fibrosis.
6. Discuss that persons with cystic fibrosis may be sterile as a result of the disease process.

CF-CM CASE MANAGEMENT

OUTCOME: The patient/family/caregiver will understand the importance of integrated case management in achieving physical and behavioral health.

STANDARDS:

1. Discuss roles and responsibilities of each member of the care team including the patient, family/caregiver, and providers in the case management plan.
2. Explain the coordination and integration of resources and services in developing and implementing the case management plan.

3. Explain the need to obtain the appropriate releases of information necessary to support integrated case management and to maintain patient privacy and confidentiality. **Refer to AF-CON**.

CF-CUL CULTURAL/SPIRITUAL ASPECTS OF HEALTH

OUTCOME: The patient/family will understand the impact and influences cultural and spiritual traditions, practices, and beliefs have on health and wellness.

STANDARDS:

1. Explain that the outcome of disease processes may be influenced by choices related to health and lifestyles, e.g., diet, exercise, sleep, stress management, hygiene, full participation to the medical plan.
2. Discuss the potential role of cultural/spiritual traditions, practices and beliefs in achieving and maintaining health and wellness.
3. Explain that traditional medicines/treatments should be reviewed with the healthcare provider to determine if there are interactions with prescribed treatment.
4. Explain that the medical treatment plan must be followed as prescribed to be effective and that some medications/treatments take time to demonstrate effectiveness.
5. Discuss that traditions, such as sweat lodges, may affect some conditions in detrimental ways. Healing customs or using a traditional healer may have a positive effect on the patient's condition.
6. Refer to clergy services, traditional healers, or other culturally appropriate resources.

CF-DP DISEASE PROCESS

OUTCOME: The patient/family will have a basic understanding of the disease process of cystic fibrosis.

STANDARDS:

1. Explain that cystic fibrosis is a genetic disorder that is autosomal recessive. This means that to have the disease, a person must inherit a gene from both parents.
2. Explain that cystic fibrosis is a chronic and progressive disease that causes mucus to become thick, dry, and sticky. This results in end organ problems especially in the lungs, pancreas, and spermatic tubules.
3. Explain that the environment, diet, exercise, or other lifestyle behaviors do not cause cystic fibrosis. The disease is not contagious and cannot be passed from one person to another except through inheritance.
4. Explain that cystic fibrosis is usually diagnosed during childhood.

5. Explain that the course of cystic fibrosis varies. Some babies show signs immediately (meconium ileus or severe respiratory problems/infections) while others may not develop symptoms for years. Some people with cystic fibrosis have a shortened life expectancy.
6. Explain the symptoms of cystic fibrosis as it applies to this patient.
7. Explain that most people with cystic fibrosis have problems with their digestive system and/or lungs. Many people have growth deficiency.
8. Explain that there is no cure for the disease but those with cystic fibrosis can live productive lives.

CF-EQ EQUIPMENT

OUTCOME: The patient/family will understand any medical equipment utilized by this patient.

STANDARDS:

1. Discuss the indications for and the benefits of prescribed medical equipment.
2. Discuss the types and features of medical equipment as appropriate.
3. Discuss and/or demonstrate proper use and care of medical equipment, participate in return demonstration by patient/family as appropriate.
4. Discuss the signs of equipment malfunction and the proper action in case of malfunction.
5. Emphasize the safe use of equipment, e.g., no smoking around O_2, use of gloves, electrical cord safety, and disposal of sharps.

CF-EX EXERCISE

OUTCOME: The patient/family/caregiver will understand the role of increased physical activity in this patient's disease process and will make a plan to increase regular activity by an agreed-upon amount.

STANDARDS:

1. Discuss medical clearance issues for physical activity.
2. Discuss the benefits of any exercise, such as improvement in well being, stress reduction, sleep, bowel regulation, and self image.
3. Assist the patient in developing a personal exercise plan. Explain that exercise should be consistent and of sufficient duration to obtain the desired outcome.
4. Encourage the patient to increase the intensity of the activity as the patient becomes more fit.
5. Discuss obstacles to a personal exercise plan and solutions to those obstacles.
6. Refer to community resources as appropriate.

CF-FU FOLLOW-UP

OUTCOME: The patient/family will understand the importance of follow-up in the treatment of cystic fibrosis.

STANDARDS:

1. Emphasize that the treatment plan and full participation to it are the responsibility of the patient/family.
2. Discuss the procedure for obtaining follow-up appointments, that follow-up appointments should be kept and the importance of continuing prescribed therapy.
3. Encourage genetic counseling prior to starting a family.
4. Discuss the availability of community resources and support services. Refer as appropriate.

CF-L LITERATURE

OUTCOME: The patient/family will receive literature about cystic fibrosis.

STANDARDS:

1. Provide patient/family with literature on cystic fibrosis.
2. Discuss the content of the literature.

CF-M MEDICATIONS

OUTCOME: The patient/family will understand the purpose, proper use, and expected outcomes of prescribed drug therapy.

STANDARDS:

1. Describe the name, strength, purpose, dosing directions, and storage of the medication.
2. Discuss the risks, benefits and common or important side effects of the medication and follow up as appropriate.
3. Discuss any significant drug/drug, drug/food, and alcohol interactions, as appropriate.
4. Discuss the importance of keeping a list of all current prescriptions and over-the-counter medicines, vitamins, herbs, traditional remedies, and supplements. Encourage the patient to bring this list and pill bottles to appointments for medication reconciliation.

CF-MNT MEDICAL NUTRITION THERAPY

OUTCOME: The patient and family will understand the specific nutritional intervention(s) needed for treatment or management of cystic fibrosis.

STANDARDS:

1. Explain that Medical Nutrition Therapy (MNT) is a systematic nutrition care process provided by a Registered Dietitian (RD) that consists of the following:
   a. Assessment of the nutrition related condition.
   b. Identification of the patient's nutritional problem.
   c. Identification of a specific nutrition intervention therapy plan.
   d. Evaluation of the patient's nutritional care outcomes.
   e. Reassessment as needed.
2. Review the basic nutrition recommendations for the treatment plan.
3. Discuss the benefits of nutrition and exercise to health and well-being.
4. Assist the patient/family in developing an appropriate nutrition care plan.
5. Refer to other providers or community resources as needed.

CF-N NUTRITION

OUTCOME: The patient/family will understand the special nutritional requirements of patients with cystic fibrosis.

STANDARDS:

1. Discuss the need for adequate calories and protein for optimal growth and development and resistance to infection.
2. Discuss as appropriate the need for pancreatic enzyme and/or salt supplementation.
3. Discuss supplementation of water miscible sources of fat soluble vitamins and iron as needed.
4. Discuss supplementation of medium chain triglyceride oils as needed.
5. Discuss the need for liberal water intake, or if extra calories are needed, calorie containing fluids. Discourage intake of dehydrating beverages such as soft drinks or other caffeinated beverages.
6. Explain that if the patient is lactose intolerant, sources of calcium other than milk may be necessary. Discuss other aspects of nutrition support as appropriate. Refer to a registered dietician for MNT or physician for specific information as appropriate.

CF-SHS SECOND HAND SMOKE

OUTCOME: The patient/family will understand the adverse health consequences associated with exposure to second-hand tobacco smoke and methods for limiting exposure of nonsmokers to tobacco smoke.

STANDARDS:

1. Define "passive smoking" and ways in which exposure occurs:
   a. Smoldering cigarette, cigar, or pipe
   b. Smoke that is exhaled from active smoker
   c. Smoke residue on clothing, upholstery, carpets or walls
2. Discuss harmful substances in smoke, e.g., nicotine, benzene, carbon monoxide, many other carcinogens (cancer causing substances).
3. Explain the increased risk of illness in the patient with cystic fibrosis when exposed to cigarette smoke either directly or via second-hand smoke.
4. Explain that cigarette smoke gets trapped in carpets, upholstery, and clothing and still increases the risk of illness even if the person with cystic fibrosis is not in the room at the time that the smoking occurs.
5. Discuss factors that increase level of exposure to second-hand smoke and preventive methods for minimizing this exposure.
6. Encourage smoking cessation or at least never smoking in the home or car. **Refer to TO-QT.**

CF-TE TESTS

OUTCOME: The patient/family will understand the tests to be performed.

STANDARDS:

1. Explain that the most common diagnostic test for cystic fibrosis is a sweat chloride test. Explain that this is a non-painful procedure.
2. Discuss the possible need for genetic testing of the patient and the impact on diagnosis and/or prognosis. Discuss the need for genetic testing for family members as well as the patient's present and future sexual partners and the impact on future progeny.
3. Explain the necessity, benefits, and risks of the test to be performed and how it relates to the course of treatment.
4. Explain the meaning of test results.

CF-TO TOBACCO (SMOKING)

OUTCOME: The patient/family will understand the dangers of smoking in the patient with cystic fibrosis and develop a plan to cut back or stop smoking.

STANDARDS:

1. Explain the increased risk of illness in the person with cystic fibrosis when exposed to cigarette smoke.

2. Encourage smoking cessation. If the patient is unwilling to stop smoking emphasize the importance of cutting back on the number of cigarettes smoked in an effort to quit or minimize increased risk of illness, hospitalization or premature death. **Refer to TO-QT.**
3. **Refer to TO**.

CF-TX TREATMENT

OUTCOME: The patient/family will understand and participate in the formulation of a treatment plan.

STANDARDS:

1. Explain that the treatment plan will be made by the patient and medical team after reviewing the available options.
2. Explain that management of cystic fibrosis varies from person to person depending on the organ systems which are involved.
3. Review the current treatment plan for this patient.
4. Discuss the importance of adhering to the treatment plan, including scheduled follow-up.